ABSTRACT

Von Hippel-Lindau (VHL) syndrome is an autosomal dominant familial neoplastic syndrome caused by mutation in VHL tumor suppressor gene localized on chromosome 3p25. The disease is characterized by abnormal vascular proliferation and increased risk of developing renal cell carcinoma, pheochromocytoma, hemangioblastoma of the central nervous system, tumors of the endolymphatic sac, cysts of the kidney, liver and pancreas, epididymal cystadenomas, neuroendocrine tumors of the pancreas, angiomas in the retina. We report a case of a bilateral pheochromocytoma, simultaneously removed by unilateral total and contralateral subtotal retroperitoneal endoscopic adrenalectomy.

Keywords: Pheochromocytoma, von Hippel-Lindau syndrome, Adrenalectomy.


Source of support: Nil

Conflict of interest: None

CASE REPORT

We present a 24-year-old woman with upper dyspeptic syndrome. On ultrasound examination, a bilateral adrenal tumor was found. Computed tomographic scan (CT) visualized a soft tissue hyperdense lesion sized 51/42 mm in the right adrenal gland as well as a similar lesion sized 41/24 mm in the left adrenal gland (Fig. 1). The tumor was functionally nonactive with normal catecholamine levels in 24-hour urine. Patient had mother with Von Hippel-Lindau (VHL) syndrome as bilateral blindness for 20 years. The genetic analysis confirmed identical mutation (p.Arg167Gln) of the VHL gene in both our patient and her mother. After appropriate preoperative assessment, total left retroperitoneal endoscopic adrenalectomy and subtotal right retroperitoneal adrenalectomy were performed in one operative time with lateral access. Four ports technique (10 mm optical port and three working—one 10 mm and two 5 mm) was used bilaterally. The first intervention was carried out on the left side in lateral decubitus position. The patient was placed with the umbilicus at the level of the break of the table to optimize trunk extension and reduce interference with instrument movement by the iliac crest. After repositioning the patient on the operating table the second stage of the operation was done. The specimen was removed with a retrieval bag. Repositioning the patient did not take more than 10 minutes. The total operative time was 180 minutes with blood loss of 100 ml, without significant intra- and postoperative complications. The patient was discharged on the third postoperative day. Histopathologically, bilateral pheochromocytoma was confirmed. There was no relapse of the disease after 5-year follow-up.

DISCUSSION

VHL syndrome is a heriditary autosomal dominant multisystem cancer syndrome. The incidence of the disease is 1:36,000.1 There is positive family history in 80% of the cases while the other 20% of the cases are due to mutation de novo.2 There is a clinical classification that divides VHL syndrome into VHL type 1—without presence of pheochromocytoma, and VHL type 2—with presence of pheochromocytoma. VHL type 2 is subdivided into type 2A (with low risk of renal cell carcinoma or cerebellar hemangioblastoma), type 2B (with high risk for developing renal cell carcinoma or cerebellar hemangioblastoma) and type 2C (familial pheochromocytoma without renal cell carcinoma or hemangioblastoma).3

Fig. 1: Computed tomographic scan of the abdomen indicates bilateral adrenal tumors
In 1988, the VHL gene on the short arm of the third chromosome was found. Later in 1993, it was cloned with establishment of its tumor suppressor activity by Latiff et al.

The VHL gene encodes a protein (pVHL), consisting of 213 amino acids and containing two functional domains—alpha and beta. The beta domain binds directly to the alpha-subunit of hypoxia-induced factor (HIF) and thus controlling the stability factor and tumor angiogenesis. The VHL type 1 syndrome is due to mutations that distort the spatial configuration of VHL protein. Ninety-six percent of the mutations responsible for VHL type 2 are missense mutations. Thus, the location, volume and type of the gene modification should predetermine phenotype of the syndrome, despite the wide variety of clinical manifestations.

For example, complete deletion of VHL gene syndrome, despite the wide variety of clinical manifestations, is not always manifested in patients with VHL syndrome. The retroperitoneal endoscopic adrenalectomy with both lateral and posterior access. The advantage of the posterior access is performance of bilateral adrenalectomy without repositioning the patient. If necessary, however, a conversion, especially in cases of profuse bleeding, is more effective in lumbal position. In bilateral intervention with lateral access repositioning the patient does not take more than 10 minutes.

Bilateral Pheochromocytoma in von Hippel-Lindau Syndrome Simultaneously Removed by Lateral Retroperitoneal

CONCLUSION
Early diagnosis, genetic consultation, adequate screening and correct therapeutic management are essential for patients with VHL syndrome. The cortex-sparing minimally invasive intervention is the contemporary therapeutic option for the treatment of pheochromocytoma in this hereditary syndrome. The retroperitoneal endoscopic adrenalectomy with lateral access is an effective and feasible method even in bilateral localization with this pathology, which however, requires extensive experience in both mini-invasive and endocrine surgery.

REFERENCES

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